Percutaneous Coronary Intervention of an Anomalous Left Circumflex Artery Originating from the Right Sinus of Valsalva: Case Report

Sağ Sinüs Valsalva’dan Köken Alan Anormal Sirkumfleks Sol Artere Perkütan Koroner Girişim

ABSTRACT
Coronary artery anomalies are rarely diagnosed during cardiac catheterization. The most common anomaly is anomalous left circumflex artery arising from the right sinus of Valsalva. In general they are asymptomatic and benign, however some patients can present with myocardial ischemia, myocardial infarction or sudden death. Treatment of atherosclerotic lesions of anomalous coronary arteries by means of percutaneous coronary intervention has rarely been reported in the literature. We present a case of a patient who was diagnosed with acute coronary syndrome and underwent an early coronary angiography followed by a successful percutaneous intervention with stenting of an anomalous left circumflex artery arising from the right sinus of Valsalva through the femoral route.

Keywords: Acute coronary syndrome; coronary vessel anomalies; angioplasty

ÖZET
Koroner arter anomalileri kalp kateterizasyonu sırasında nadiren teşhis edilmektedir. En sık görülen koroner anomali sağ sinüs Valsalva’dan köken alan anormal sol sirkumfleks arterdir. Koroner arter anomalileri genellikle asemptomatik ve benign seyirli olmalarına rağmen bazı hastalarda miyokard iskemisi, miyokard infarktüsü ve ani ölüm ile sonuçlanabilmektedir. Literaturede anormal koroner arterlerin aterosklerotik lezyonlarının tedavisinde perkütan koroner girişim na
diren bildirilmştir. Bu makalede akut koroner sendrom tanısıyla femoral yoldan erken koroner anjiyografi yapılan ve sağ sinüs Valsalva’dan kaynaklanan anormal sol sirkumfleks artere başarılı perkütan girişim yapılan bir olguya sunmaktayız.

Anahtar Kelimeler: Akut koroner sendrom; koroner damar anomalileri; anjiyoplasti

C oronary artery anomalies (CAAs) are incidentally found in 0.6-1.5% of all coronary angiographies (CAGs). A higher incidence of coronary anomalies is observed in young victims of sudden death than in adults (4-15% vs. 1%, respectively). In a large retrospective incidence study, CAAs were found in 1% of adults and in 0.9% of children. Although CAAs are often asymptomatic, life-threatening symptoms such as; myocardial ischemia and infarction, cardiac arrhythmias, syncope and sudden death can occur especially during exercise. Anomalous origin of a coronary artery from the pulmonary artery is one of the
most serious anomalies, with a 90% mortality rate during infancy period. After hypertrophic cardiomyopathy, CAAs are the second most common cause of sudden death in young athletes. Anomalous coronary arteries comprise a major challenge during diagnostic procedure but recognition and adequate visualization is essential, especially in patients undergoing evaluation for percutaneous coronary intervention (PCI), coronary artery surgery or prosthetic valve replacement.

The most common coronary anomaly, firstly described by Antopol and Kugel in 1933 and reported to have prevalence of 0.18-0.67%; is anomalous left circumflex artery (ALCx) arising from the right sinus of Valsalva through a common or separate ostium along with the right coronary artery (RCA) or directly as a branch of the RCA itself.3-8 It is usually considered to be benign since it is not known to predispose individuals to sudden cardiac death.9,10 However, some have proposed an increase in coronary artery disease (CAD) in these aberrant vessels due to acute angulation of its origin from the aorta.11 We present a case of an 87-year-old male patient who was diagnosed with acute coronary syndrome (ACS) and underwent a successful PCI with stenting of ALCx arising from the right sinus of Valsalva through the femoral route.

CASE REPORT

The patient provided a written informed consent with regard to this report. An 87-year-old hypertensive male was diagnosed with ACS and referred to our clinic for CAG. On admission, blood pressure, pulse rate, respiratory rate and body temperature were 100/60 mmHg, 92 beats per minute, 20 per minute and 36.6 °C, respectively. Cardiac and pulmonary examinations were normal. Electrocardiogram revealed minimal ST segment depressions in the anterior leads. Chest radiography was normal. Transthoracic echocardiography revealed a regional hypokinesia in the inferolateral segments of the left ventricle with an estimated left ventricular ejection fraction of 50%. Laboratory tests were normal except an elevated level of troponin T (0.111-reference 0.000-0.025 ng/mL). GRACE score at admission was 172, which indicates an ACS at high risk of adverse cardiac events. Medical therapy including ticagrelor, aspirin, angiotensin-converting enzyme, beta-blocker, statin, nitrate and heparin was initially implemented. An early invasive approach was reasonable for this patient. CAG revealed atherosclerotic plaques in left anterior descending and right coronary arteries and significant lesions in proximal, mid and distal segments of an ALCx which was arising from a separate ostium located in close relationship with the right aortic sinus (Figures 1, 2 and Videos 1, 2). The patient was planned for PCI of both proximal and mid lesions of the ALCx. The distal lesion was not suitable for PCI because of small diameter. The ALCx was selectively cannulated with 6F Mach1 left guiding catheter (Boston Scientific). Then the lesions were crossed with two 0.014”×182 cm ChoICE wires (Boston Scientific) for adequate support and a 2.75×18 mm BioMatrix Flex stent (Biosensors Interventional) was deployed at 14 atm at the proximal lesion (Video 3). Pre-dilatation with 1.5×15 mm and 2.0×15 mm

FIGURE 1: Coronary angiography demonstrating the anomalous left circumflex artery arising from the right sinus of Valsalva with consecutive atherosclerotic lesions.
Invader PTCA balloon dilatation catheters (Alvimedica) at 18 atm and 14 atm respectively was performed at the mid ALCx lesion. A 2.5×24 mm BioMatrix Flex stent (Biosensors Interventional) was deployed at 14 atm (Video 4). There was no need for post-dilatation. There was no complication at the end of the intervention (Figure 3 and Video 5). The patient was followed up with medical therapy and was discharged with ticagrelor (90 mg BID), aspirin (100 mg/day), atorvastatin (40 mg/day), metoprolol succinate (50 mg/day) and ramipril (10 mg/day) on the third day without any adverse cardiac event.

**DISCUSSION**

Left circumflex artery (LCx) originating from the right sinus of Valsalva is a well-known anatomical variation.\(^1\)\(^6\) The first case of PCI performed for such aberrant vessels was described in 1982.\(^12\) During CAG two angiographic signs have been previously described and have been proven to be reliable in recognizing the anomalous artery before selective demonstration. The “aortic root sign”, defined as a profile view of the artery behind the aortic root during left ventriculography, and the “non-perfused myocardium sign”, described as a recognition of absent arterial inflow to a significant area of the posterior lateral left ventricle during selective injections of the left main coronary artery. In our case selective opacification of the left main coronary artery revealed an absent LCx and engaging the ALCx was not difficult. Therefore, we didn’t perform a left ventriculography or an aortography.

Despite being classified as a benign anomaly, angina pectoris, myocardial ischemia, myocardial infarction and sudden death have been reported even in the absence of atherosclerotic lesions.\(^11\)\(^13\)-\(^14\) These manifestations might be due to repeated compression of the anomalous artery by a dilated aortic root or to unusual angling of the LCx, which can compress the coronary ostium and cause ischemia. Although some studies have demonstrated ALCx to be with no increased incidence of atherosclerosis, our case, demonstrating significant atherosclerotic lesions only in ALCx, may support that acute angulation, potential retroaortic course and increased shear stress of ALCx may contribute in the development of atherosclerosis in these vessels.

During diagnostic CAG attention should especially be given to rare CAAs. The condition when a coronary artery appears to be as an extension of another coronary artery is a seldom and confusing anomaly. Recently Demirkol et al. reported a case of LCx appears as a terminal extension of RCA.\(^15\) In such circumstances, operator should be aware of such anomalies to avoid
longer procedure time and more radiation exposure. It is frequently difficult to selectively cannulate the anomalous coronary artery. The failure to adequately engage the anomalous vessel may lead to poor angiographic visualization and an erroneous diagnosis. Likewise, lack of guide support may lead to PCI failure. Based on available literature, when cannulation of the ALCx and aligning catheter during PCI comprise challenge amplatz, coronary bypass or multipurpose guide catheters can be considered to get adequate support. If engagement of the vessel is still insufficient catheter can be quietly slid over the guide wire while the predilatation balloon is still inflated. In our case, it was difficult to get the predilatation balloon through the culprit lesion of the ALCx. We overpassed this challenge by using two guide wires to align the vessel and get more support. In addition, jailed wire technique made it easier for us to pass the second stent through the stent which had been deployed in the proximal part of the vessel.

In summary, CAAs are rare and should be suspected especially in young adults presenting with cardiac symptoms. Although, PCI of anomalous vessel can be inconvenient, intervention can be managed by using different guide catheters and techniques. We hereby describe a case of atherosclerotic ALCX artery arising from the right sinus of Valsalva which was successfully treated by PCI. As there are no established treatment guidelines, revascularization would be considered only in those patients with significant atherosclerotic changes and documented ischemia. To the best of our knowledge, this case report is one of few similar cases described in the literature.

Conflicts of Interest
There are no conflicts of interest.

Authorship Contributions:

Idea/concept: Constructing the hypothesis or idea of research and/or article: Alaa Quisi; Design: Planning methodology to reach the conclusions: Alaa Quisi; Control/supervision: Organizing, supervising the course of progress and taking the responsibility of the research/study: Mustafa Gür; Data Collection and/or Processing: Taking responsibility in patient follow-up, collection of relevant biological materials, data management and reporting, execution of the experiments: Alaa Quisi; Analysis and/or Interpretation: Taking responsibility in logical interpretation and conclusion of the results: Alaa Quisi; Literature Review: Taking responsibility in necessary literature review for the study: Alaa Quisi; Writing the Article: Taking responsibility in the writing of the whole or important parts of the study: Alaa Quisi, Mustafa Gür; Critical Review: Reviewing the article before submission scientifically besides spelling and grammar: Mustafa Gür; References and Fundings: Providing personnel, environment, financial support tools that are vital for the study: Mustafa Gür; Materials: Biological materials, taking responsibility of the referred patients: None.

REFERENCES


Video 1: Coronary angiography demonstrating the anomalous left circumflex artery arising from the right sinus of Valsalva with consecutive significant atherosclerotic lesions.

Video 2: Coronary angiography demonstrating significant atherosclerotic lesions of the anomalous left circumflex artery arising from the right sinus of Valsalva.

Video 3: Coronary angiography demonstrating stent deployment at the proximal lesion of the anomalous left circumflex artery.

Video 4: Coronary angiography demonstrating stent deployment at the mid lesion of the anomalous left circumflex artery.

Video 5: Coronary angiography demonstrating the final result after stent deployment at the proximal and mid lesions of the anomalous left circumflex artery with TIMI 3 flow.